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Letter to the Editor

Oral and cutaneous findings are valuable diagnostic aids in Wegener's granulomatosis

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Wegener's granulomatosis (WG) is a rare granulomatous necrotizing vasculitis of small vessels which has predilection for upper airways, lung and kidney, but can involve any other organ. Without treatment, the mainstay of which is the combination of cyclophosphamide and systemic corticosteroids, it may run a fatal course. In a previous issue, Ruokonen et al. have reported a case presenting with oral lesions as the sole sign of WG [1]. In the abovementioned case, the patient had hyperplastic granular gingivitis called as strawberry gingivitis which is a rare manifestation of WG, but may be the first sign and nearly pathognomonic for this autoimmune vasculitis in around 5% of patients with WG [2]. Interestingly, also the histopathological features of the so-called strawberry gingivitis, albeit nonspecific, are very suggestive of WG and can be seen in only few other conditions affecting the gingiva: they are characterized by the combination of pseudoepitheliomatous hyperplasia, epithelial microabscesses, and intense inflammatory infiltrate of neutrophils, eosinophils and multinucleated giant cells [3]. It is noteworthy that the differential diagnosis of WG based on oral manifestations is wide and includes most notably other vasculitis such as microscopic polyangiitis, specific granulomatous diseases such as sarcoidosis and Crohn's disease, granulomatous infectious processes such as tuberculosis and deep mycoses, blood dyscrasias such as leukemia and lymphoma, and drug-induced gingival changes. As dermatologists, in addition to the differential diagnoses mentioned above, we would highlight a rare condition, namely Langerhans cell histiocytosis (LCH), which is a multisystem disorder characterized by the proliferation of a distinct cell type — the so-called Langerhans cell — and presenting with distinctive mucocutaneous manifestations. In LCH, vegetating nodulo-ulcerative lesions closely resembling those of WG may involve the gums and be accompanied by tooth mobility as in strawberry gingivitis [4,5]. As reported by Ruokonen et al., it has been suggested that oral manifestations as well as cutaneous findings are valuable diagnostic aids in WG. In fact, about 15% of patients with WG develop specific skin lesions at some time during the course of the disease or more rarely at disease onset [6]. The most common skin lesion is palpable purpura with the histopathological correlate of leukocytoclastic vasculitis. Nodules, papules, ulcerations and deep erythema nodosum-like subcutaneous nodules complete the wide clinical spectrum. In conclusion, the case on Ruokonen et al. emphasizes the importance to recognize the oral and cutaneous manifestations of WG, which may also precede systemic involvement, to start proper treatment as soon as possible so improving the outcome of this potentially lethal disease.

References

[1] Ruokonen H, et al. "Strawberry like" gingivitis being the first sign of Wegener's granulomatosis. Eur J Intern Med 2009;20(6):651–3.
[2] Patten SF, Tomecki KJ. Wegener's granulomatosis: cutaneous and oral mucosal disease. J Am Acad Dermatol 1993;28:710–8.
[3] Lourenco SV, Nico MM. Strawberry gingivitis: an isolated manifestation of Wegener's granulomatosis? Acta Derm Venereol 2006;86(1):90–1.
[4] Caputo R, Marzano AV, Passoni E, Fassati LR, Agnelli F. Sclerosing cholangitis and liver transplantation in Langerhans cell histiocytosis: a 14-year follow-up. Dermatology 2004;209(4):335–7.
[5] Stewart C, Cohen D, Bhattacharyya I, Scheitler L, Riley S, Calamia K, et al. Oral manifestations of Wegener's granulomatosis: a report of three cases and a literature review. J Am Dent Assoc 2007;138(3):338–48.
[6] Comfere NI, Macaron NC, Gibson LE. Cutaneous manifestations of Wegener's granulomatosis: a clinicopathologic study of 17 patients and correlation to antineutrophil cytoplasmic antibody status. J Cutan Pathol 2007;34(10):739–47.

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